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Case Series

Schwannomas at unusual locations: a case series

Sumitava Pachal*, Sudipta Chatterjee, Supratim Chaki

Department of General Surgery, Midnapore Medical College and Hospital, Paschim Medinipur, West Bengal, India

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***Correspondence:**
Dr. Sumitava Pachal,
E-mail: sumitavapachal@ymail.com

ABSTRACT

Schwannoma, also known as neurinoma or neurilemmoma, is a benign, encapsulated, slow-growing, peripheral nerve sheath tumor arising from the Schwann cells. They are seen mostly in the head and neck region, with the 8th cranial nerve being the most common site. There it is known as vestibular Schwannoma or acoustic neuroma. It is usually seen in 20–50 years of age, with no specific sex or racial predilection. Histopathologically, there may be compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Nuclear palisading around fibrillary processes, also known as Verocay bodies, is often seen in cellular areas. In this article, we present a series of cases where all of the patients presented to us with soft tissue swelling for varying periods of time in different locations, mostly in extremities. Cytopathology and radiology couldn't come to a specific diagnosis. The tumors were excised en mass. The post-operative histology of the excised tumor confirmed the diagnosis of Schwannoma, arising from peripheral nerve sheath. There was no recurrence after follow up for 1 year. Schwannomas most commonly arise from the peripheral nerve sheaths usually at the head neck region. They are seen rarely in other peripheral nerves. They should be excised en mass with intact epineurium. After complete excision their recurrence rate is very low thus having a good prognosis.

Keywords: Neurilemmoma, Peripheral nerve sheath tumor, Schwannoma

INTRODUCTION

Schwannoma is a benign, peripheral nerve sheath tumor arising from the Schwann cells. They are mostly seen in adults 20–50 years of age, majorly in the head-neck region.[1] The most common location is the 8th cranial nerve, where it is popularly known as acoustic neuroma or vestibular Schwannoma. It can affect any peripheral nerve, but the incidence rate is rare, being 0.6 in 100,000 populations.[2] Sciatica is characterized by pain along the back, buttock, thigh, leg, and foot caused by irritation or compression of one of the 5 spinal nerve roots or nerve trunk of the sciatic nerve. In 1% of cases, sciatica is associated with Schwannoma of the sciatic nerve.[1,3]

As it is a slow-growing tumor, and the symptoms often attributed to lumbosacral degenerative pathology, the diagnosis is often delayed for a longer period of time.[2,4,5]

CASE SERIES

A total of 8 patients, attending the general surgery outpatient department, between June 2022 to October 2022, who were primarily diagnosed as soft tissue tumor, involving mostly the extremity, were taken up for thorough history taking and clinical examination. There was mostly a female preponderance, with a mean age of the subjects being 43 years. All of them came with the common complaint of a firm to hard swelling in the extremity for a variable duration. One female patient had the swelling located in the abdomen, in the left iliac fossa. Dull aching pain was a common complain, although there was no significant history of numbness of the area distal to it or paresthesia, or any sensory or motor neurodeficit. Grossly, the swellings, on examination were well-circumscribed, having a firm-to-harm consistency, non-compressible, non-fluctuant, and having a smooth surface with regular

margins, free from the overlying skin but attached to the underlying structures.

All of the subjects underwent series of investigation after proper history and clinical examination. Hematological parameters were checked alongside radiological investigation defining the tumor characteristics. In most of the cases, ultrasonography (USG) described the lesions to be heterogeneously hypoechoic SOL. In few cases, internal vascularity was also noted. Magnetic resonance imaging (MRI) was done to delineate soft tissue. The female who had the left iliac fossa mass, underwent fine needle aspiration cytology (FNAC) and contrast enhanced computerized tomographic (CECT) scan of the lower abdomen. On FNAC, it was said to be a spindle cell neoplasm. On CECT whole abdomen, the lesion was said to be homogenous with no intraperitoneal extension.

After proper optimization of the parameters, the subjects were planned for en mass removal of the tumor. During excision, the tumors were found to be arising from peripheral nerve sheath, namely sciatic nerve (in the 1st case), common peroneal nerve (in the 2nd case), lateral cutaneous nerve of calf (in the 3rd case), cutaneous nerve of anterior abdominal wall (in the 4th case), cutaneous nerves of the arm and forearm (in the 5th and 6th case respectively), radial nerve (in 7th and 8th case respectively). Post-operative outcome was uneventful in all of the cases. Post-operative histopathological examination revealed that they were Schwannomas, having hyper-cellular (Antoni A) areas with nuclear palisading pattern, and relatively hypo-cellular (Antoni B) area with myxoid degeneration. All of them were followed up for 1 year, where there was no recurrence. A brief summary of all the cases with post-op complications have been described in Table 1.

Results and interpretation

There was a female preponderance in this study with Schwannomas occurring in the extremities were mostly seen. Mean age of presentation was 1 and a half years. Mean post-operative stay was 3 days and on follow up, recurrence was not seen in any of the cases in 1-year duration.

Our patients had Schwannomas involving different peripheral nerves. Extremities were most commonly involved than trunk. A careful *en mass* excision was done in all of the cases. The histopathology of the specimen confirmed the diagnosis of Schwannoma or neurilemmoma. During the entire post-operative period, the patients didn't develop any neurodeficit. After 1 year, during follow-up, none of them reported any recurrence.

Table 1: Demographic trend with post-operative outcome of the subjects.

S. no	Age (years)	Sex	Duration	USG	MRI	Post-operative stay	Recur-rence	Post-op complicat-ions
1	50	M	8 years	Heterogeneous hypoechoic SOL	PNS tumour	5	Nil	Nil
2	56	F	10 years	Heterogeneous hypoechoic lesion	Heterogeneous intensity s/o neoplastic aetiology	3	Nil	Pain, serous discharge
3	21	F	1 and half years	Heterogeneous lesion with internal vascularity	Soft tissue sarcoma - neurogenic tumour	3	Nil	Nil
4	45	F	9-10 months	Solid and heterogeneous lesion	FNAC - spindle cell neoplasm. CECT W/A -homogenous lesion with no intraperitoneal extension	4	Nil	Pain
5	35	F	2 and half years	Heterogeneous SOL	Soft tissue tumour - sarcoma	2	Nil	Nil
6	42	F	1 and half years	Heterogeneous hypoechoic lesion with internal vascularity	PNS tumour	3	Nil	Nil
7	45	M	2 years	Heterogeneous hypoechoic SOL	Soft tissue sarcoma – neurogenic tumour	3	Nil	Serous discharge
8	51	M	1 and half years	Heterogeneous lesion with internal vascularity	PNS tumour	3	Nil	Nil

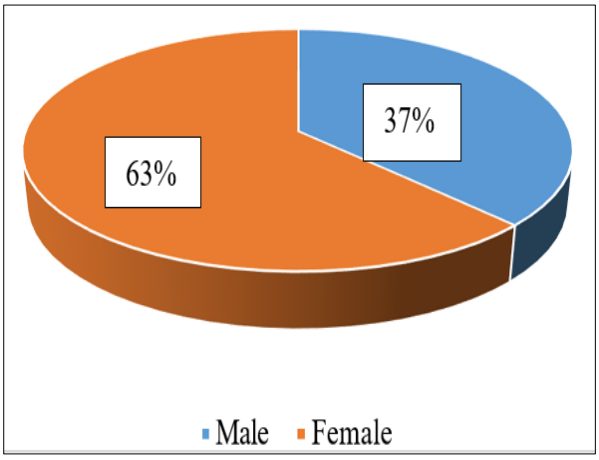

Figure 1: Distribution of study subjects as per sex.

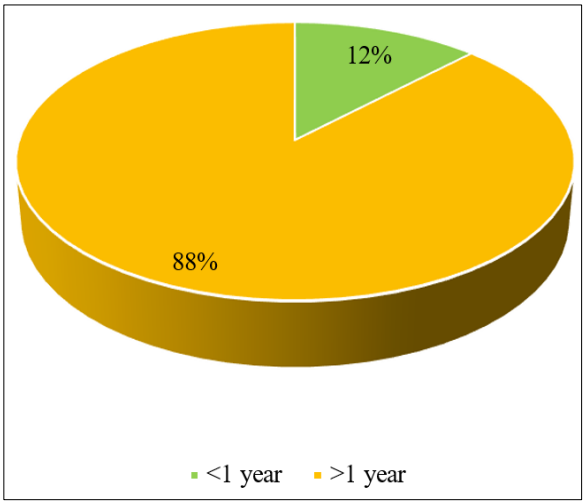

Figure 2: Distribution of study subjects as per duration of symptoms.

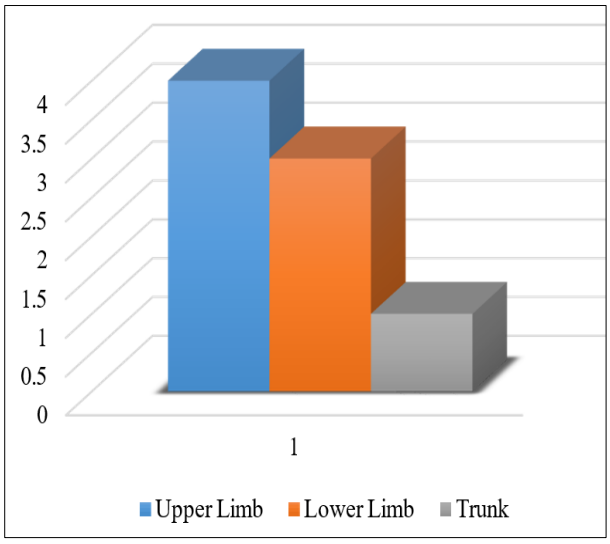

Figure 3: Distribution of study subjects as per anatomical location of the tumor.

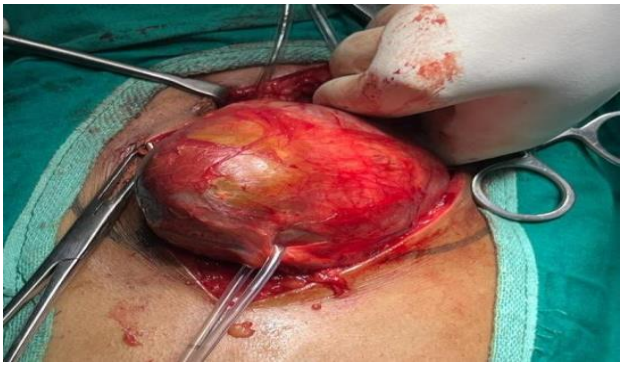

Figure 4: Sciatic Schwannoma intraoperatively.

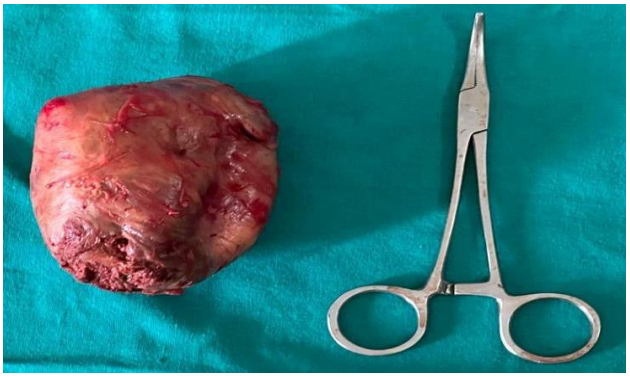

Figure 5: Schwannoma after excision en mass.

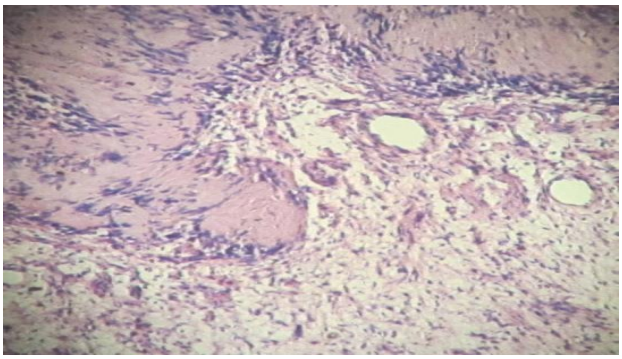

Figure 6: Histology of Schwannoma.

DISCUSSION

Schwannoma, also known as neurinoma of Verocay, neurilemmoma, is a benign, encapsulated, slow- growing peripheral nerve sheath tumor that originates from the differentiated Schwann cells of the neuroectoderm. These Schwann cells form the myelin sheath that insulates the nerves and facilitates impulse transmission.[6] In about 90% of cases, they are sporadic. Very few cases have been seen to have syndromic associations (neurofibroma type 2, Schwannomatosis, and Carney complex).[7-10] Of all nerve sheath tumors, Schwannoma is the most common type, comprising almost 89% of the cases. They generally affect the main trunk of the nerve, more specifically in the upper limb, followed by the head, trunk, and flexor surfaces of the lower extremities. The most common site of benign

Schwannoma is the 8th cranial nerve, where it is also known as acoustic neuroma or vestibular Schwannoma. It comprises of almost 60%.[11] Schwannomas are usually seen in adults 20–50 years of age.[1] But there is no sex or racial predilection. The incidence of sciatic Schwannoma is extremely rare, comprising 0.6 per 100,000 populations.[2] The incidence of Schwannoma of the other peripheral nerves, namely the superficial peroneal or the lateral cutaneous nerve of calf, as we had found them to be, or in the anterior abdominal wall, is even rarer.[22-27]

Schwannomas are slow-growing tumors and may often be asymptomatic sometimes. They can present in various locations with a lot of variable clinical manifestations. Tenderness is often felt while palpating the mass. There may also be neurologic symptoms if the tumor is large. In the extremities, they may present as an asymptomatic mass, with mild pain in the area, or paresthesia due to pressure on the parent nerve. Schwannoma of the sciatic nerve can be asymptomatic or can present with pain, paresthesia, or neurological deficit. In very few cases (~1%), it can be associated with sciatica.[1,3]

Plain radiography is usually non-specific. Hence, magnetic resonance imaging (MRI) and computed tomography (CT) scans are among the special studies to consider. CT scan is not as sensitive or specific for the diagnosis of Schwannoma as MRI but is often the first investigation obtained. It is particularly useful in assessing bony changes adjacent to the tumor. The MRI usually shows an oval or round mass with an isointense or hypo-intense signal on T1-weighted images and a hyper-intense, heterogeneous signal on T2-weighted images.[12] The lesion is enhanced uniformly with gadolinium contrast. Split-fat sign (the thin peripheral rim of fat seen on planes along the long axis of the lesion in non-fat-suppressed sequences), Target sign (central area of hypo-intensity with a peripheral T2 signal hyper-intensity), or fascicular sign (multiple small ring-like structures) may also be seen.

The differential diagnosis of Schwannoma includes neurofibroma, malignant peripheral nerve sheath tumor, leiomyoma or leiomyosarcoma, chordoma, chondroblastoma, and malignant melanoma. Histopathologically, there are different varieties of Schwannoma including the classic Schwannoma (encapsulated tumor having hypercellular Antoni A areas with spindle cells palisading around eosinophilic areas, known as Verocay bodies, and hypocellular with myxoid degeneration Antoni B areas), cellular Schwannoma (composed of entirely Antoni A areas and is devoid of Verocay bodies), plexiform Schwannoma (plexiform growth having an Antoni A pattern) or the melanotic Schwannoma (having epithelioid cells and melanin accumulated in neoplastic cells and melanophages).

Schwannomas respond positively to local resection. They can be excised en mass, as they arise within the nerve sheath and are surrounded by a true capsule comprised of epineurium, enabling a complete enucleation without damage to the parent nerve.[2,3,13,18,19] If the surgeon suspects that complete resection would cause a permanent neurologic deficit, they might call for an intra-lesional resection or stereotactic radiosurgery.[14,15] Postoperative complication is significantly higher in younger patients, with large tumor size, deep location of the tumor, and tumors originating from the ulnar nerve.[21] Frequently reported complications include neuropraxia, vascular complications, and injury to the main nerve trunk.

The prognosis of Schwannoma is excellent. Recurrence after total resection is very uncommon. The risk of malignant transformation is approximately 18% in neurofibromatosis type 1, and 5% in Schwannomas.[3,16-17] The prognosis depends on the size, location of the tumor, and underlying conditions.[20]

CONCLUSION

Schwannomas are mostly benign entity. If excised properly en mass, then their prognosis is very good as the rate of recurrence is very low.

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REFERENCES

1. Kralick F, Koenigsberg R. Sciatica in a patient with unusual peripheral nerve sheath tumors. Surg Neurol. 2006;66:634-7.
2. Nawabi DH, Sinisi M. Schwannoma of the posterior tibial nerve: the problem of delay in diagnosis. J Bone Joint Surg Br. 2007;89:814-6.
3. Rekha A, Ravi A. Sciatic nerve schwannoma. Int J Low Extrem Wounds. 2004;3:165-7.
4. Ghaly RF. A posterior tibial nerve neurilemoma unrecognised for 10 years: case report. Neurosurgery. 2001;48:668-72.
5. Blanchard C, Dam-Hieu P, Zagnoli F, Bellard S. Chronic sciatic pain caused by sciatic nerve schwannoma. Rev Med Intern. 2008;29:748-50.
6. Rustagi T, Badve S, Parekh AN. Sciatica from a foraminal lumbar root schwannoma: case report and review of literature. Case Rep Orthop. 2012;2012:142143.
7. Lee SU, Bae YJ, Kim HJ, Choi JY, Song JJ, Choi BY, et al. Intralabyrinthine Schwannoma: Distinct Features for Differential Diagnosis. Front Neurol. 2019;10:750.
8. Coy S, Rashid R, Stemmer-Rachamimov A, Santagata S. Correction to: An update on the CNS manifestations of neurofibromatosis type 2. Acta Neuropathol. 2020;139(4):667.
9. Ahlawat S, Blakeley JO, Langmead S, Belzberg AJ, Fayad LM. Current status and recommendations for imaging in neurofibromatosis type 1, neurofibromatosis type 2, and schwannomatosis. Skeletal Radiol. 2020;49(2):199-219.

10. Vindhyal MR, Elshimy G, Elhomsy G. StatPearls. StatPearls Publishing; Treasure Island (FL). 2021.
11. Propp JM, McCarthy BJ, Davis FG, Preston-Martin S. Descriptive epidemiology of vestibular schwannomas. Neuro Oncol. 2006;8(1):1-11.
12. Skolnik AD, Loevner LA, Sampathu DM. Cranial Nerve Schwannomas: Diagnostic Imaging Approach. Radiographics. 2016;36(5):150199.
13. Calisaneller T, Gok H, Yilmaz MS, Cinar GK, Naderi S. Schwannoma of sciatic nerve: a rare cause of non-discogenic sciatica. World Spinal Column J. 2011;2:77-80.
14. Park HH, Hong CK, Jung HH, Chang WS, Kim CH, Lee WS, et al. The Role of Radiosurgery in the Management of Benign Head and Neck Tumors. World Neurosurg. 2016;87:116-23.
15. Sade B, Mohr G, Dufour JJ. Vascular complications of vestibular schwannoma surgery: a comparison of the suboccipital retrosigmoid and translabyrinthine approaches. J Neurosurg. 2006;105(2):200-4.
16. Topsakal C, Akdemir I, Tiftikci M, Ozercan I, Aydin Y. Malignant schwannoma of the sciatic nerve originating in a spinal plexiform neurofibroma associated with neurofibromatosis type 1—case report. Neurol Med Chir (Tokyo). 2001;41:551-5.
17. Rezzouk J, Durandeau A. Compression nerveuse par pseudokyste mucoïde: à propos de 23 cas. Rev Chir Orthop Reparatrice Appar Mot. 2004;90:143-6.
18. Lamond RC, Fox B. Management of peripheral nerve tumors. Limb Preservation. 2004;11.
19. Thiebot J, Laissy JR, Delangre T, Biga N, Liotard A. Benign solitary neurinomas of the sciatic popliteal nerves CT study. Neuroradiology. 1991;33:186-8.
20. Kano H, Kondziolka D, Khan A, Flickinger JC, Lunsford LD. Predictors of hearing preservation after stereotactic radiosurgery for acoustic neuroma. J Neurosurg. 2009;111(4):863-73.
21. Hirai T, Kobayashi H, Akiyama T, Okuma T, Oka H, Shinoda Y, et al. Predictive factors for complications after surgical treatment for schwannomas of the extremities. BMC Musculoskelet Disord. 2019;20(1):166.
22. Shariq O, Radha S, Konan S. Common peroneal nerve Schwannoma: an unusual differential for a symptomatic knee lump. BMJ Case Reports. 2012;bcr2012007346.
23. Dane B, Dane C, Basaran S, Erginbas M, Cetin A. Vaginal Schwannoma in a case with uterine myoma. Ann Diagn Pathol. 2010;14:137-9.
24. Liu Y, Chen X, Wang T, Wang Z. Imaging observations of a schwannoma of low malignant potential in the anterior abdominal wall: A case report. Oncol Lett. 2014;8:1159-62.
25. Mishra A, Hamadto M, Azzabi M, Elfagieh M. Abdominal wall schwannoma: case report and review of the literature. Case Rep Radiol. 2013;2013:456863.
26. Tarchouli M, Essarghini M, Qamouss O, El Kharras A, Bounaim A. Abdominal wall schwannoma: a case report. Gastroenterol Hepatol Bed Bench. 2020;13(1):95-100.
27. Nascimento G, Nomi T, Marques R, Leiria J, Silva C, Periquito J. Ancient Schwannoma of superficial peroneal nerve presenting as intermittent leg pain: A case report. Int J Surg Case Rep. 2015;6C:19-22.

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